Case Report

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A Rare Report of Vaginal Rhabdomyosarcoma

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Abstract

Sarcomas are connective tissue cancers and Rhabdomyosarcomas are cancers originating from the skeletal muscle cells of the body. A 23 year old female unmarried, nulligravida presented in our emergency with the complaint of bleeding per vagina on and off since last 4 months. The lesion measured 9.2 (cc) x8.7 (cs)x7.8 (ap) cm. The lesion displaced the uterus superiorly distended the vaginal canal compressed the urinary bladder and urethra anteriorly but with no evidence of infiltration into the pelvic side wall and with maintained fat plane. Rest all pelvic organs as well as abdominal organs were within normal limit. The HPE report on microscopic examination of the tissue showed features suggestive of malignant spindle cell neoplasm. Immunohistochemistry was done and IHC report showed: Positive for Desmin, Myogenin predominantly cytoplasmic positive. Correlating the morphologic and IHC markers the report came to be Embryonal Rhabdomyosarcoma (Botryoid Rhabdomyosarcoma). As soon as the final diagnosis was made patient was referred to Department of Oncology and chemotherapy with Cyclophosphamide was started. She has been on regular follow up since then and is keeping well. The presence of a cervical polyp in an adolescent is a gynecologic oddity and must necessarily be examined histologically because it might be a rhabdomyosarcoma. This is extremely important because diagnosis at an early stage of the disease is a highly favorable prognostic factor.

Keywords: Vaginal Rhabdomyosarcoma; Bleeding per vagina; Mesenchymal cells.

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INTRODUCTION

Sarcomas are connective tissue cancers and Rhabdomyosarcomas are cancers originating from the skeletal muscle cells of the body. They originate from the rhabdomyoblasts (mesenchymal cells) and hence are tumors presenting in the children and adolescent age group.[1] The common sites are head and neck (35%), urogenital tract (22%), extremities (18%) and trunk.[1,2] In adults the soft tissue sarcomas constitute less than 1% of all malignancies and out of that 3% are rhabdomyosarcomas.[2] Most of the cases of adult rhabdomyosarcomas have

been reported as case reports and case series. According to WHO Rhabdomyosarcomas can be broadly classified into 4 histologic subtypes, - Embryonal, Alveolar, Pleomorphic, Spindle cell/ Sclerosing rhabdomyosarcomas.[3] Botryoid rhabdomyosarcoma is a polypoid variant of embryonal rhabdomyosarcoma and constitutes approximately 3% of all rhabdomyosarcomas.[4] In active reproductive age group the primary site of occurrence is the cervix.[5]

CASE REPORT

A 23 year old female unmarried, nulligravida presented in our emergency with the complaint of bleeding per vagina on and off since last 4 months with no definite pattern of flow or duration or regularity. The abnormal bleeding was associated with mild lower pain abdomen and difficulty in passing urine since last 6 months, along with occasional retention of urine for which she required catheterisation.

She attained menarche at 15 years and all her previous cycles were regular till last 4 months.

She had no significant medical or surgical illness in the past.

The patient had sought medical help in a nearby Government Maternity hospital in the month of March, 2021 for retention of urine and difficulty in micturition and was referred to the Department of Urology, NEIGRIHMS for the same on out-patient basis. Patient was evaluated for the presenting complaint and a CECT abdomen was done which revealed a well defined heterogeneously enhancing soft tissue lesion, in the centre of the vagina. The lesion measured 9.2 (cc) x8.7 (cs)x7.8 (ap) cm. The lesion displaced the uterus superiorly distended the vaginal canal compressed the urinary bladder and urethra anteriorly but with no evidence of infiltration into the pelvic side wall and with maintained fat plane. Rest all pelvic organs as well as abdominal organs were within normal limit.

Later, the patient again presented to Government Maternity hospital in view of recurrent abnormal uterine bleeding. She underwent the cervical growth biopsy and the HPE report showed intermediate to high grade spindle cells with the possibilities of

- Cellular/ mitotically active leiomyoma
- Intermediate/high grade endometrial stromal sarcoma.

A few days later the patient again presented to the Government Maternity hospital with the complaints of heavy bleeding per vagina for continuous 16 days, and due to haemodynamic instability and clinical pallor patient was admitted in the said institute. Patient was transfused with 4 units of Packed red blood cells and CE-MRI was done. The MR pelvis showed large (16.1 x 8.6 x 13.5 cms) lobulated cystic mass with locoregional extension with aggressive angiomyoma of the vagina.

The patient then attended our hospital and on clinical examination patient was found conscious oriented and haemodynamically stable. No clinical pallor seen. On per abdomen examination a solid firm central mass of around 22 weeks size uterus was found with restricted mobility, non-tender, lower margin not palpable.

On per speculum examination a fleshy mass which bled on touch, filling the entire of introitus was seen. Cervix couldn't be visualised due to the mass. On gentle per vaginal examination cervical os couldn't be reached due the growth. While performing per vaginum examination due to friability of the growth tissue was obtained and sent for HPE.

USG whole abdomen done stated that a heterogeneously solid lesion centered in the cervical stroma with echogenic thickened ectocervix can be seen with vascularity and mass effect, probable stromal sarcoma of ectocervix.

The HPE report on microscopic examination of the tissue showed features suggestive of malignant spindle cell neoplasm. Immunohistochemistry was done and IHC report showed: Positive for Desmin, Myogenin predominantly cytoplasmic positive, SMA negative – which ruled out leiomyosarcoma, CD 10 negative – which ruled out endometrial stromal sarcoma, Pan CK negative – which ruled out carcinoma. Correlating the morphologic and IHC markers the report came to be Embryonal Rhabdomyosarcoma (Botryoid Rhabdomyosarcoma).

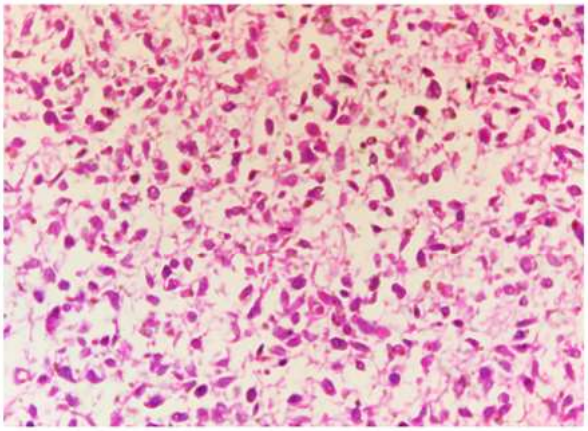

Fig. 1: Photomicrograph showing sheets of stellate, spindled and strap cells in a myxoid background (H & E, x400)

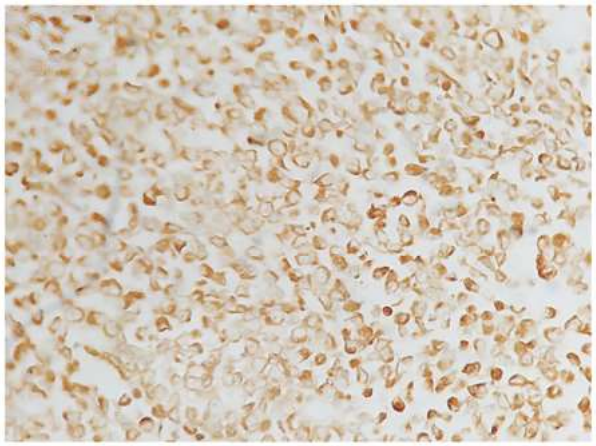

Fig. 2: Tumour cells showing positivity for desmin (IHC, x400)

As soon as the final diagnosis was made patient was referred to Department of Oncology and chemotherapy with Cyclophosphamide was started. She has been on regular follow up since then and is keeping well.

DISCUSSION

Embryonal rhabdomyosarcomas are rare cancers of the cervix, occurring in less than 0.2% of all cervical malignancies.[6,7] Embryonal rhabdomyosarcomas usually affect adolescent age group and young adults.[7] The classic presentation of cervical rhabdomyosarcoma is abnormal bleeding and a palpable mass in the vagina.[6] The standard treatment remains controversial, because of the rare occurrence and presentation and individualised management options according to the treating team, such as, radical surgery, adjuvant therapies. Historically the prognosis of embryonal rhabdomyosarcoma is poor, however with adjuvant multidrug chemotherapy with Vincristine, Actinomycin D, Cyclophosphamide combined with local radiation therapy and/or surgery has increased the survival rates.[8] Irradiation therapy should be considered for patients with positive lymph nodes.[8] In our patient fortunately there was no palpable lymph nodes. Some case reports in the literature have mentioned the occurrence of multiple primary cancers in patients with cervical rhabdomyosarcomas notably Sertoli-Leydig cell tumors.[9-12] Rhabdomyosarcomas in general have been associated with certain inherited diseases, such as Beck with Wiedemann syndrome, Li-Fraumeni syndrome, neurofibromatosis type 1, Noonan syndrome, Costello syndrome MEN2A syndrome.[13] According to the staging of Rhabdomyosarcoma in general, a tumour arising in the genital area other than bladder or prostate is Stage 1 and falls in low-risk group if PAX/FOX 01 gene fusion is absent.[14] The survival rates are highest in the pre-pubertal and adolescent girls, where the disease incidence is also high.[15] Older age group, non-embryonal histologic type and advanced stage of disease has inferior outcomes.[15]

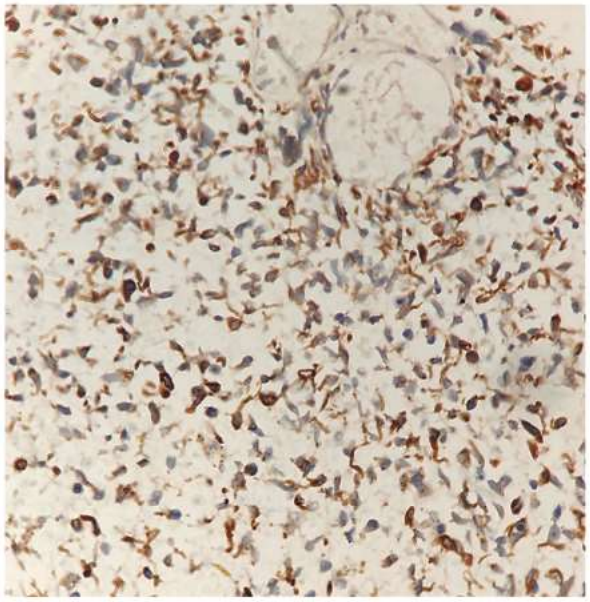

Fig. 3: Tumour cells showing positivity for myogenin (IHC, x400)

CONCLUSION

The presence of a cervical polyp in an adolescent is a gynecologic oddity and must necessarily be examined histologically because it might be a rhabdomyosarcoma. This is extremely important because diagnosis at an early stage of the disease is a highly favorable prognostic factor.

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